

# Acral pseudolymphomatous angiokeratoma – a case report<sup>\*</sup>

## *Angioqueratoma pseudolinfomatoso acral – relato de caso<sup>\*</sup>*

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**Abstract:** A 29-year-old female patient with asymptomatic red-brown papules that had arisen four years before in her right forearm. Two lesions were removed for histopathological and immunohistochemical exams. Intralesional corticosteroid injection resulted in no improvement and the lesions were subsequently surgically excised with complete resolution of the condition.

**Keywords:** Immunohistochemistry; Pseudolymphoma; Skin

**Resumo:** Paciente de 29 anos, do sexo feminino, com aparecimento há quatro anos de pápulas assintomáticas, eritemato-acastanhadas, no antebraço direito. Duas das lesões foram biopsiadas para exames histopatológico e imuno-histoquímico. A paciente foi inicialmente tratada com corticoterapia intralesional, sem melhora. Procedeu-se, então, à exérese das lesões com resolução completa do quadro.

**Palavras-chave:** Imuno-histoquímica; Pele; Pseudolinfoma

### INTRODUCTION

Acral pseudolymphomatous angiokeratoma is a benign subcutaneous tissue condition that, in spite of histopathological similarities with lymphomatous processes, must be included in the group of pseudolymphomas of the skin. It was described by Ramsay in 1988, after the observation of five children (one male and four females) who presented multiple violet erythematous papules, located unilaterally, in the foot in four cases, and in the hand in one case, thereby the denomination acral. It is known by its acronym Apache – Acral Pseudolymphomatous Angiokeratoma of Children.<sup>1,3</sup>

Apache etiology remains unknown; however, some authors believe in the possibility that it is a hypersensitivity reaction to insect bites, in virtue of the histopathological picture and acral location of the lesions.<sup>3,5</sup>

It is clinically characterized by the presence of 10 to 40 violet-erythematous papules, whose diameter

ranges from 1 to 4 mm, and which are asymptomatic, located unilaterally, generally with acral distribution and occurring more often between 2 and 13 years of age. It should be distinguished upon histopathological examination from the following clinically similar diseases: Mibelli's angiokeratoma, hemangioma, melanocytic nevus, basocellular carcinoma, lymphocytoma cutis and cutaneous lymphocytoma.<sup>1,2</sup>

Histological studies of Apache reveal an epidermis of normal aspect and a dense well-differentiated lymphocyte infiltrate in the dermis, amongst connective tissue structures, not affecting skin annexes. Such histopathological aspect configures lymphocyte proliferation, albeit with no nuclear atypies, which requires an immunohistochemical study in order to differentiate from lymphomas.<sup>1,3</sup>

Recommended therapies for Apache include total exeresis of skin lesions, intralesional steroids or radiation therapy.<sup>2,3</sup>

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### CASE REPORT

Twenty-nine-year-old brow-skinned female patient, who presented with an asymptomatic sessile brownish-erythematic papule in the right forearm, measuring approximately 3mm in diameter, four years ago. One year ago, the cutaneous condition evolved with the onset of other four lesions, with similar pattern, next to the first (Figure 1). She underwent two biopsies, and histopathological examinations revealed an epidermis with normal aspect, dense lymphocyte infiltrate amongst dermal connective tissue structures, and no compromising of skin annexes, which configures nodular lymphocyte proliferation (Figure 2), thus demanding differential diagnosis with lymphoproliferating diseases, by means of Immunohistochemistry. The patient did not present lesions in any other region of the skin, nor systemically.

Immunohistochemical study of the biopsy material revealed positivity for B-lymphocytes (CD20+), T-lymphocytes (CD3+) and light chains of immunoglobulins *kappa* and *lambda* in the plasma cell population (Figures 3, 4, 5 ad 6), yielding the conclusion, from the combination of histopathological and immunohistochemical findings, that this was a polyclonal leukocyte infiltrate (B and T cells), indicating an inespecific reactional inflammatory infiltrate, with no evidence of malignancy, which then excluded the possibility of lymphoma.

An Intralesional infiltration with triamcinolone was performed, with no efficacy, as shown by immediate relapse of the lesions. Choice was then made for total excision of the lesions, with complete resolution of the dermatological process until after a year of follow-up.

### DISCUSSION

Acral pseudolymphomatous angiokeratoma exhibits clinical similarities with Mibelli's angiokeratomas and with hemangiomas. Histopathologically, lymphocyte proliferation with no nuclear atypies is observed, characterizing a "pseudolymphoma", whose main differential diagnoses are lichen nitidus, lymphocytoma cutis, bowenoid papulosis and reaction to insect bite. Performance of an immunohistochemical study following inclusion in paraffin is recommended for exclusion of cutaneous lymphoma proper, since, in acral pseudolymphomatous angiokeratoma, lymphocyte infiltrate consists of both B and T cells. Occasionally, histiocytes and plasma cells are found among the lymphocytes, which confirms the reactional character of the process, unlike cutaneous lymphoma proper, where only T or B cells proliferate.<sup>1-5</sup>

As mentioned, even though the acronym Apache denotes acral location and higher frequency in children, this disease is not exclusively located in the extremities, nor is it restricted to that age range.<sup>1</sup> Okada et al.,<sup>6</sup> when describing three clinical cases in females aged 11, 41 and 52 years with clinical lesions similar to those of Apache, proposed the term acral angiokeratoma-like pseudolymphoma. Such denomination was due to the clinical aspect, which was similar to angiokeratoma, albeit with histopathological features of pseudolymphoma. Ohtsuka and Yamazaki<sup>7</sup> described the case of a 28-year-old female patient and also referred to clinical diagnosis as acral angiokeratoma-like pseudolymphoma. Besides being painful, intralesional steroid injection proved to be ineffective for treating the patient's cutaneous lesions, and, since radiation therapy is a very aggressive method to



FIGURE 1: Asymptomatic papules in the right forearm, measuring approximately 3 mm in diameter

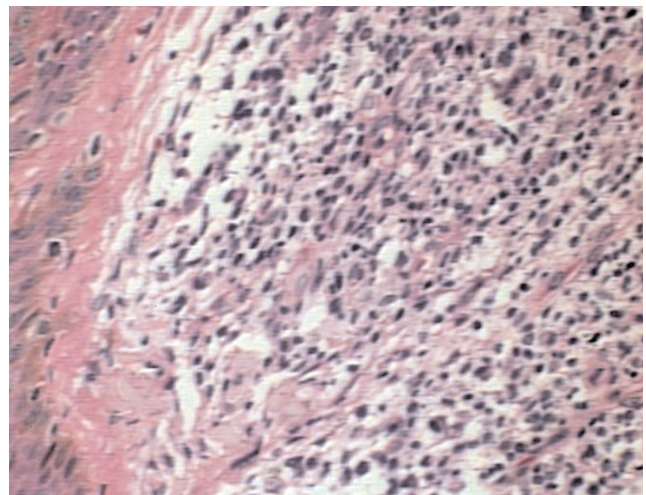


FIGURE 2: Leukocyte infiltrate with predominance of lymphocytes (HE x 40)



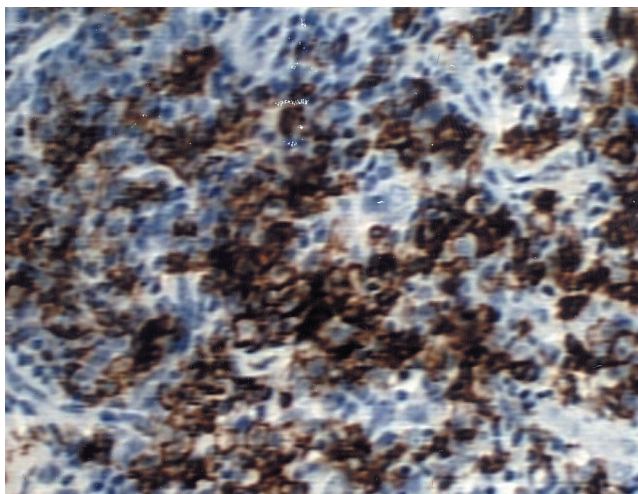


FIGURE 3: Immunohistochemistry: CD20 – positive, in dark brown, in B-lymphocytes (HE x 40)

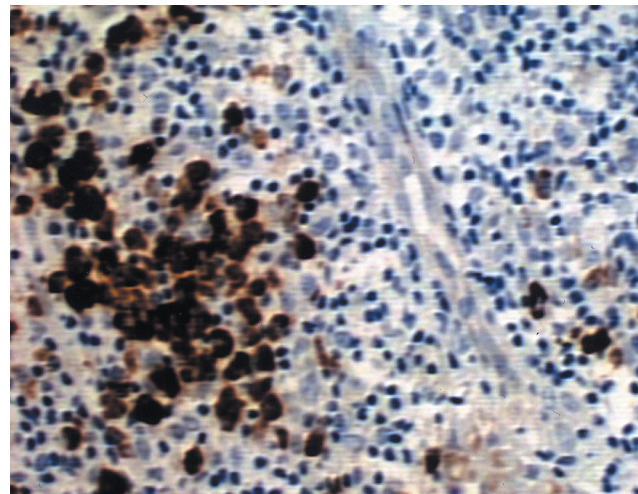


FIGURE 5: Immunohistochemistry: light chain of kappa-positive immunoglobulin, in brown (HE x 40)

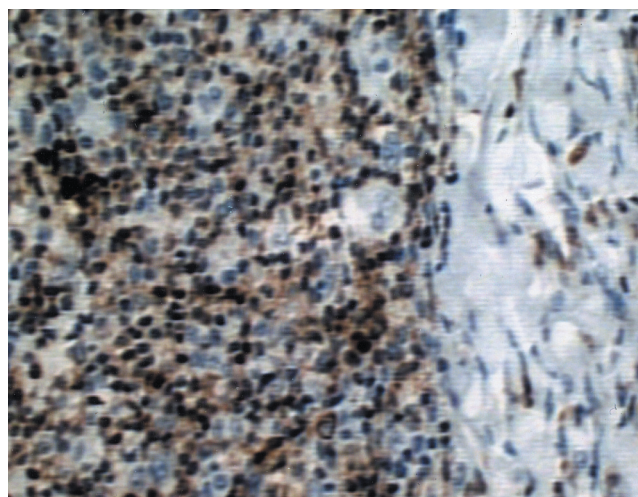


FIGURE 4: Immunohistochemistry: CD3 – complex associated with T-positive lymphocyte receptors, in dark brown (HE x 40)

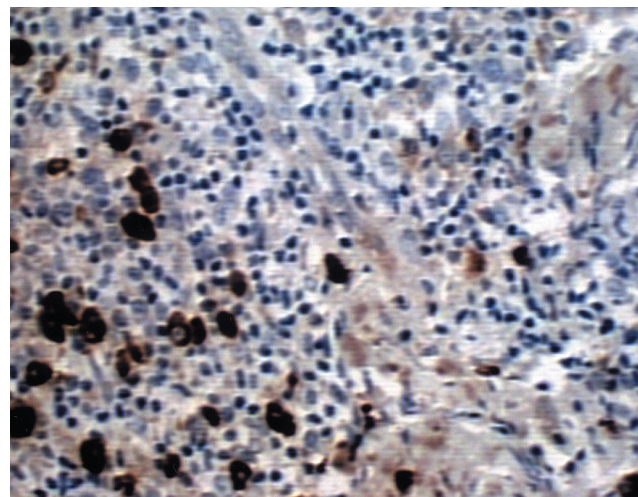


FIGURE 6: Immunohistochemistry: light chain of lambda-positive immunoglobulin, in brown (HE x 40)

be used upon such a benign condition, the authors chose to carry out a total excision of the lesion, which was eventually the best therapeutic option, as the patient is still cured after one year of follow-up.<sup>3</sup>

In brief, acral pseudolymphomatous angiokeratoma is a rare, benign, cutaneous pseudolymphoma, characterized by the presence of unilateral violet-

erythematic papules, usually acral and often between the ages of 2 and 13 years, which requires both histopathological and immunohistochemical evaluations, in order to make differential diagnosis with cutaneous lymphomas. Although uncommon, there is a possibility of occurrence in adults and outside extremities, as observed in the case reported here.<sup>1-5</sup> □

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